

Fig 2A

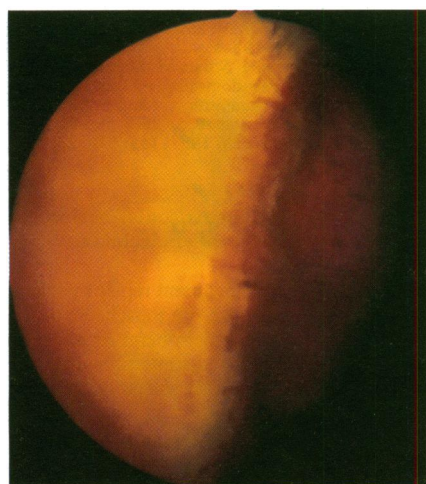


Fig 2B

Figure 2 Fundal photographs showing (A) the neovascular complex (NV) arising anterior to the indent, which regressed (B) after cutting the encircling band.

On examination, his left visual acuity was perception of light and a hyphaema and vitreous haemorrhage prevented any fundal view. The eye was normotensive and B-scan ultrasound revealed no abnormality other than the scleral indent. Two weeks after presentation, the haemorrhage had cleared sufficiently to reveal the presence of rubeosis iridis (Fig 1A) and a large neovascular complex arising from the retina at 9 o'clock anterior to the circumferential indent (Fig 2A), confirmed by anterior segment and fundal fluorescein angiography. The retina was normal posterior to the indent. Although initially normotensive, the intraocular pressure was 46 mm Hg 1 month later. A mild anterior uveitis also developed. The encircling band was cut but not removed under general anaesthesia, and the intraocular pressure was 20 mm Hg 1 day postoperatively. The retinal indent was much shallower. On review 2 weeks postoperatively the eye was not hyperaemic, intraocular pressure was 15 mm Hg, and by 1 month postoperatively the rubeosis and anterior retinal neovascularisation had resolved (Fig 1B and 2B). His visual acuity remains 6/18 with +11.00/-1.25x160 aphakic correction.

COMMENT

The patient suffered a vitreous haemorrhage owing to the development of retinal neovascularisation arising anterior to the scleral indent with normal retina posteriorly. We believe that he had chronic ischaemia of the retinal periphery and anterior segment,

induced by the right encircling band, possibly predisposed by his systemic hypertension and long history of cigarette smoking.

Anterior segment ischaemia is a rare but recognised complication of scleral buckling surgery, with a reported incidence of 3% in a general retinal detachment population¹ but is far more common (71%) in patients with sickle SC disease.² It presents typically soon after surgery with corneal oedema, anterior chamber flare, and ocular hypotension. The ischaemia may be due to either impaired blood supply to³ or venous return from⁴ the anterior segment. A tight encircling band might impair both the long posterior ciliary arteries, which contribute to the choroidal circulation, and the choroidal venous drainage. The anterior ciliary arteries may also be compressed by an explant placed under the rectus muscles. These vessels and the vortex veins may also be damaged preoperatively by diathermy, cryotherapy, or detachment of two or more rectus muscles.

However, the onset of ischaemic signs in the present case was later than is typical for anterior segment ischaemia, and rubeosis iridis is also atypical. Ocular ischaemic syndrome, which may occur in older men and result in neovascular glaucoma and visual loss, is also a possibility. Carotid and retinal Doppler ultrasound studies were obtained only postoperatively in this patient, but showed no abnormality. This case has many similarities with anterior hyaloidal fibrovascular proliferation described in diabetics following vitrectomy and cataract surgery.⁵ This has only been reported in diabetics, and usually those with active proliferative retinopathy. Diabetes mellitus was excluded in this patient, however, and the 1 year interval between vitrectomy and the neovascularisation is longer than would be expected.

The causative role for the band in the ischaemia in this case is strongly suggested by the distribution of the retinal neovascularisation anteriorly but not posteriorly, and the rapid regression of both the irideal and retinal neovascularisation once the band was removed; increased retinal artery blood flow has been demonstrated following removal of encircling elements.⁶ Retinal neovascularisation is an unusual complication of scleral encirclement surgery.

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Peter's anomaly in an infant with a mild degree of Axenfeld's anomaly

EDITOR,—We treated a 5-month-old boy with type II Peter's anomaly associated with a mild degree of Axenfeld's anomaly. He had several iris strands attached to the posterior embryotoxon of the right eye, microcorneas, and lateral displacement of the inner canthi of both eyes. To our knowledge, only four cases of Peter's anomaly with Axenfeld's anomaly have been reported, so this case is probably the fifth report of Peter's anomaly.¹⁻⁴

CASE REPORT

The patient was a 5-month-old male who had been delivered spontaneously at full term. There was no family history of ocular diseases. At birth bilateral corneal opacities were noted. At 5 months he was referred to our hospital for examination. The inner canthi were displaced laterally (Fig 1). The inter-outer canthi distance/inter-inner canthi distance was 2.0 (normal range 2.45-2.96). Slit-lamp biomicroscopy showed central opacities in both eyes, the anterior chamber

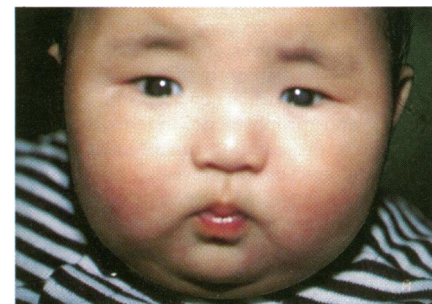


Figure 1 The face of the patient. Exotropia of the left eye and lateral displacement of the inner canthi are observed.

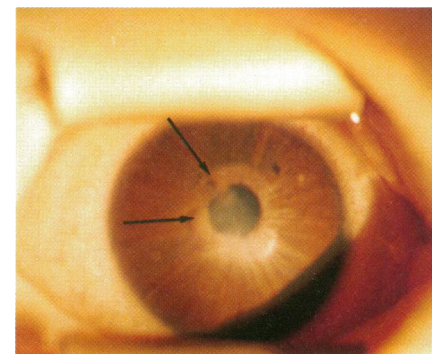


Figure 2A Anterior segment of the right eye. Microcornea, 9 mm in diameter, with small sharply demarcated central corneal opacity and anterior synechiae at 9 o'clock and 11 o'clock (arrows) are apparent.



Figure 2B Slit-lamp examination of the right eye after mydriasis. Iris-corneal adhesions are visible.

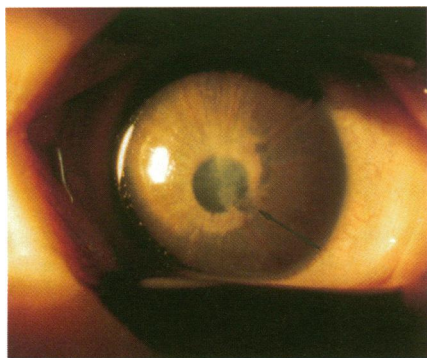


Figure 3 Anterior segment of the left eye. Microcornea, 6 mm in diameter, with a small sharply demarcated central opacity and an anterior synechia except at 4-5 o'clock (arrow) are noted.

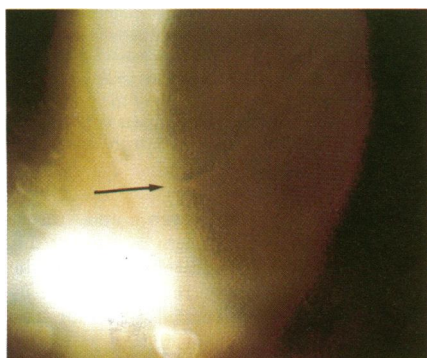


Figure 4 Gonioscopic finding with gonioprism of the right eye. There is an iris strand to the temporal posterior embryotoxon (arrow).

was shallow, and the central iris was attached to the periphery of the corneal opacity.

We diagnosed Peter's anomaly. Haematological analysis and chromosomal analysis with G-banding were all within the normal range. A paediatrician found no developmental retardation or systemic malformation. Hypertelorism was excluded. Ultrasonography showed a normal posterior segment.

Intraocular pressure was 12 mm Hg (RE) and 13 mm Hg (LE) with a portable applanation tonometer under general anaesthesia. The corneal diameters were 9 mm (RE) and 8 mm (LE) (that is, microcorneas). In the right eye, anterior synechia were present at 9 o'clock and 11 o'clock near the margin of the central corneal opacity (Figs 1A and B). In the left eye, an anterior synechia was present at the margin of the central corneal opacity except at 4-5 o'clock (Fig 3). Gonioscopic examination with a prism revealed several iris strands attached to the temporal and upper portions of a prominent Schwarbe's line, posterior embryotoxon in the right eye (Fig 4). In the left anterior chamber angle, we identified posterior embryotoxon but no iris strand was identified. Ophthalmoscopy showed a normal right optic disc and intact retina. The ocular fundus of the left eye was hardly visible because of corneal opacity. Ophthalmic examinations of his parents revealed no abnormal findings. There was no evidence of chromosomal abnormality of them. At 15 months, the ocular tension had been within the normal range.

COMMENT

Most cases of Peter's anomaly are sporadic, although reports of parental consanguinity and more than one affected sibling support an autosomal recessive or irregularly dominant mode of inheritance in some cases. Our

patient is sporadic. He has several iris strands attached to the posterior embryotoxon in the temporal and upper parts of the anterior chamber of the right eye. This patient also has a mild degree of Axenfeld's anomaly without glaucoma.

Mesenchymal cells which differentiate into corneal endothelium, stroma, iris, and aqueous outflow structure have been proved to be derived from neural crest cells by histochemical method.^{5,6} Bahn *et al*⁷ suggested that corneal endothelial disorders, Peter's anomaly, congenital glaucoma, posterior embryotoxon, Axenfeld's anomaly, Rieger's anomaly, and sclerocornea result from abnormal neural crest cell migration. Thus, in most cases of Peter's anomaly and of Axenfeld's anomaly the pathogenesis is related to abnormal migration of neural crest cells. It is speculated that the pathogenesis in our patient with Peter's and Axenfeld's anomalies is related to abnormal migration of the neural crest cells. If this hypothesis is correct, it would seem that the already reported cases of Peter's anomaly with Axenfeld's anomaly are disproportionately scarce. The reason for this disproportion may be that: (1) the wide corneal opacity conceals the anterior chamber; (2) most patients with this anomaly are babies or children, so gonioscopic examination is very difficult without general anaesthesia; (3) in patients with synechia between the central iris and the margin of the corneal opacity, the anterior chamber angle is hard to visualise, because of the central iris obstruction, with a gonioscopic mirror without a gonioscopic prism.

Careful gonioscopic examination with a prism under general anaesthesia of patients with Peter's anomaly would presumably reveal latent Axenfeld's anomaly.

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Subretinal hypopyon in polymphocytic leukaemia

EDITOR,—It is well known that leukaemias can manifest themselves in ocular tissues. Most clinical and pathological studies suggest an incidence of at least 50% among leukaemic patients.¹ Ocular involvement is

markedly more frequent in acute than in chronic disease. All ocular tissues may become involved, mostly, however, choroid and retina. Retinal manifestations are haemorrhages, exudates, congested tortuous vessels, perivascular sheathing, and exudative or haemorrhagic retinal detachment which are summarised under the term leukaemic retinopathy. We report on a clinical sign of leukaemic retinopathy which has not been described previously, a subretinal hypopyon.

CASE REPORT

A 59-year-old male patient suffered from polymphocytic leukaemia for 6 years. After splenectomy which had been performed 1 year after onset the leukaemic state was stable with a white cell count between 100 and 150×10⁹/l, no anaemia, and no thrombocytopenia. For 9 months the disease progressed showing an increasing white cell count. Chemotherapy had to be administered. When the patient was referred to our department he complained of blurred vision and ocular pain. At that time white cell count was 400×10⁹/l, haemoglobin 8.0 mg/100 ml, and platelets 200×10⁹/l. Visual acuity was 0.8

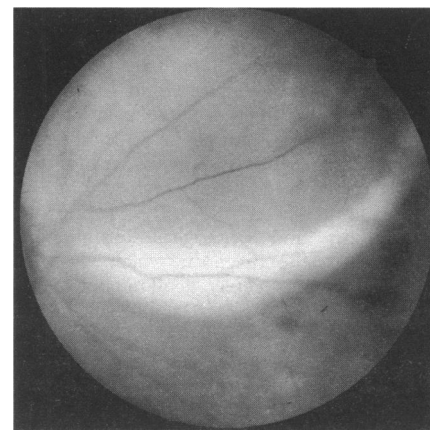


Figure 1 Subretinal aggregation of leukaemic cells at the bottom of serous retinal detachment in leukaemic retinopathy.

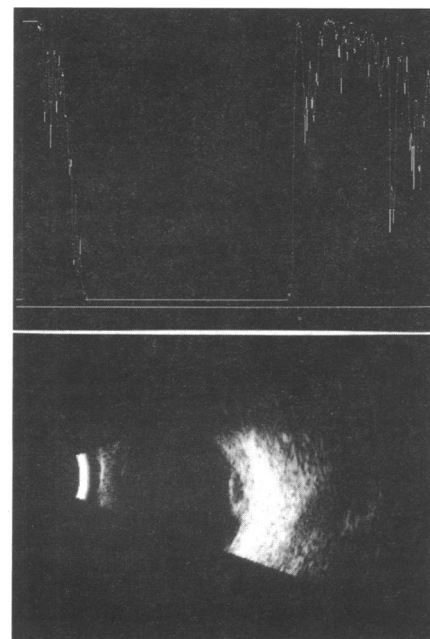


Figure 2 A- and B-scan in upright position: aggregation of cellular elements at the bottom of subretinal serous fluid (for further explanation see text).